the 3M/House implant for children, mirroring the widespread professional acceptance of such devices in children. The 3M device is no longer available, but the Nucleus multichannel cochlear implant received FDA marketing approval for use in children in June 1990.

Extensive auditory, speech, educational, and psychological testing is done before and after implantation for children. Results show that cochlear implants provide auditory detection over much of the speech signal. Compared with before implantation, there is considerable improvement in auditory discrimination and speech production skills. Limited open-set word and sentence recognition is possible for at least some children, perhaps as many as 30%. Complications with the device have been minimal but include inappropriate placement of the electrode array, postauricular flap infection, facial paresis, transient dizziness or pain, and partial device extrusion. About 1% of patients in the Nucleus clinical trials had unresolved complications.

Selection criteria include bilateral profound sensorineural deafness, a lack of substantial benefit from hearing aids, psychological suitability, and medical suitability. The primary criterion for selection is confirmation of profound sensorineural hearing loss, usually with an average unaided threshold for the speech frequencies of poorer than 110 dB or an aided speech detection threshold of poorer than about 53 dB. Patients who are candidates show poorer performance on selected auditory discrimination tasks while using appropriately fit hearing aids than those using an implant. Families and, if possible, candidates should be well motivated and possess appropriate expectations. Medical contraindications include deafness caused by lesions of the acoustic nerve or central auditory pathway, active middle ear infections, cochlear ossification that prevents electrode insertion, an absence of cochlear development, and tympanic membrane perforation.

The cochlear implant can provide sound to deaf adults and children unable to benefit from hearing aids. The complex assessment, rehabilitation, and counseling should be done by centers with the multidisciplinary staff necessary to provide effective care for patients with this special auditory prosthesis.

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Anticytoplasmic Autoantibody Test for Wegener's Granulomatosis

WEGENER'S GRANULOMATOSIS is a systemic disease characterized by necrotizing laryngeal granulomas with vasculitis of the upper and lower respiratory tract, systemic vasculitis, and focal necrotizing glomerulitis. The disease can be limited or generalized. When there are nasal, pulmonary, and renal findings, a clinical diagnosis and histologic confirmation by nasal or kidney biopsy are relatively simple.

In Wegener's disease with limited and localized findings, such as isolated laryngeal subglottic involvement, diagnosis may be difficult. Not every nasal or laryngeal biopsy specimen will demonstrate characteristic histologic findings. The finding of a nonspecific inflammatory infiltrate is more likely.

The anticytoplasmic autoantibody test has been shown to have high specificity for Wegener's granulomatosis. The test was also useful to monitor reactivation and remissions of the disease. The absence of the autoantibody does not rule out Wegener's disease. The test is useful in diagnosing the disease in patients in whom histologic confirmation is not always possible—such as those with isolated subglottic stenosis. The test is done with a standard immunofluorescence technique. The cytopreparation is considered positive for anticytoplasmic autoantibody if the characteristic focal, centrally accentuated, fine granular cytoplasmic staining pattern is present in most neutrophils. Experience with pattern recognition and strict adherence to the methodologic protocol are essential for obtaining reproducible and reliable results.

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Gadolinium Magnetic Resonance Imaging in Bell's Palsy

IT HAS BEEN DISCOVERED that the facial nerve in Bell's palsy has a characteristic appearance on gadolinium-enhanced magnetic resonance imaging (Gd-MRI) scans. Despite this important advance in imaging technology, the clinical presentation remains the mainstay of diagnosis. The diagnosis of Bell's palsy in the past has been one of exclusion, arrived at when a rapid onset of facial weakness is unaccompanied by signs or symptoms suggestive of disease involving the central nervous system, temporal bone, or parotid gland. More than 80 causes have been identified that result in facial paralysis. Many patients with serious underlying diseases affecting the facial nerve, such as tumors and infections, have been inappropriately given the diagnosis of Bell's palsy. Such a misdiagnosis may delay needed therapy and diminish the probability of eventual nerve recovery. When a facial palsy has atypical features, the diagnosis of Bell's palsy should be questioned, and a Gd-MRI scan of the nerve obtained. The most common reason for doing a Gd-MRI scan is weakness that persists for a longer period than would be expected in an uncomplicated Bell's palsy. When paralysis persists beyond two months, the diagnosis of idiopathic paralysis should be questioned. Other atypical features include slowly progressive palsy, facial palsy accompanied by spasm, recurrent palsy, unusual degrees of pain, and the presence of multiple cranial neuropathies or other neurologic symptoms.

On Gd-MRI scans, the facial nerve in Bell's palsy appears bright because of gadolinium uptake in the inflamed segment. This is usually most pronounced in the region of the geniculate ganglion at the lateral end of the internal auditory canal, but more diffuse enhancement of the intratemporal portion may be seen. This enhancement typically persists for several months and may not resolve entirely, even after paral-

ysis has abated. The Gd-MRI appearance tends to confirm the theory that Bell's palsy results from viral mononeuritis. Swelling of the nerve within the tight confines of the fallopian canal in the temporal bone results in ischemia and eventual neural dysfunction. The diameter of the nerve is the key difference between Bell's palsy and tumors involving the nerve. In Bell's palsy, the nerve is bright but not swollen or otherwise distorted, whereas in tumors, the nerve is typically both bright and focally enlarged. Gadolinium-MRI scans of patients with facial palsy should include the entire course of the facial nerve, from its exit from the brain stem to its terminal branches on the face.

In the recent past, clinicians managing a patient with atypical or persistent facial palsy faced a dilemma. If imaging studies were normal, the cause could be either atypical Bell's palsy or a subtle anatomic lesion beyond the resolving capacity of computed tomography or an unenhanced MRI study. The ability to make a positive diagnosis of Bell's palsy with Gd-MRI is reassuring to patients and clinicians. Indeed, many patients with Bell's palsy fear they have had a stroke, and an MRI scan may occasionally be warranted to allay those fears. A possible use for Gd-MRI in Bell's palsy stems from the preliminary observation that the degree of gadolinium enhancement correlates with the prognosis for the recovery of facial function. When intense gadolinium uptake involves a long segment of the nerve, a prolonged clinical course and incomplete recovery appear more probable.

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Laser Therapy for Early Cancer of the Vocal Cords

"EARLY," SUPERFICIAL squamous cell cancer, limited to mucous membrane of the membranous portion of the vocal cord, can be treated with an 80% to 90% chance of cure with irradiation or surgical therapy. Surgical resection in the past was by cup forceps removal (stripping) of the mucosa of the membranous vocal cord. It is now done more precisely with a dry field using the laser through microdirect laryngoscopy.

For laser (or cup forceps) stripping to be successful, the cancer should not invade more than 1 to 2 mm below the basement membrane of the mucosa, and the best results, of course, are obtained when it is a squamous cell carcinoma in situ.

Normal vocal cord mobility is an indication that there is no invasion into Reinke's space just below the mucosa, much less into the thyroarytenoid muscle. This is confirmed with a suction tube tip by palpation of the involved vocal cord through direct laryngoscopy. The abnormal mucosa is then excised with a 1-mm normal mucosal margin and a thin layer of normal connective tissue under the lesion. The lesion needs to stop at least 1 mm from the anterior commissure, its lateral edge should be visible on the floor of the ventricle (thus not go up into the false vocal cord), it should not extend onto the body and preferably not onto the vocal process of the arytenoid, and it should not go more than 3 to 4 mm below the free edge of the vocal cord. The margins should be clear and sharp.

The disadvantage of laser treatment is that irradiation achieves an equal five-year survival rate of about 90% and is equally effective when there is some invasion or spread slightly onto the arytenoid or onto the anterior opposite vocal cord or when margins are not sharply delineated. Although radiation therapy takes six weeks and can leave dryness in the throat, its main advantage is that it achieves a better voice on average than vocal cord stripping, even when the latter is done by an experienced laser surgeon through microdirect laryngoscopy. In summary, irradiation is advised as the primary therapy for T_1 and $T_2N_0M_0$ vocal cord squamous cell carcinoma. In carefully selected patients, however, especially after irradiation has failed, and if the lesion still meets (and originally met) the described criteria, laser resection of the lesion with a small margin around and beneath it may be a good alternative to vertical hemilaryngectomy or total laryngectomy.

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Autoimmune or Immune-mediated Causes of Deafness

CERTAIN FORMS OF rapidly progressive sensorineural hearing loss have an autoimmune or immune-mediated basis. Patients with these forms commonly have bilateral ear involvement that begins as fluctuating hearing loss but becomes progressive and often profound. There is an accompanying disequilibrium or outright ataxia if the vestibular system is also affected. About 20% to 30% have associated, definable manifestations of systemic autoimmune disease, such as systemic lupus erythematosus, polyarteritis nodosa, or ulcerative colitis. Diagnosis is difficult because no certain means exists to identify this disorder. The use of lymphocyte transformation against inner ear antigens has led to different results. Recent studies using a Western blot immunoassay against crude inner ear antigen have shown some diagnostic promise; of 54 patients with rapidly progressive hearing loss, 19 were positive on Western blot, but only 1 of the 14 (7%) normal persons showed a similar band.

Other means of evaluating this disorder include a sedimentation rate, the antinuclear antibody test, and C1q binding and Raji cell assays. These nonorgan-specific tests help identify a generalized autoimmune state in a patient.

Treatment consists of a trial of high-dose steroids—prednisone, 60 mg daily for three to four weeks. Improved hearing will often be evident after three weeks. Maintenance on alternate-day therapy should be attempted with as low a dose as possible, as long as hearing is maintained. Some investigators recommend giving cyclophosphamide—2 to 5 mg per kg of body weight a day—in patients who do not respond to steroids and who are not of childbearing age, as the drug has potential oncogenic risks.